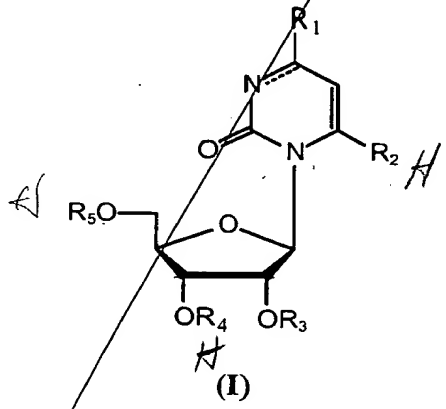
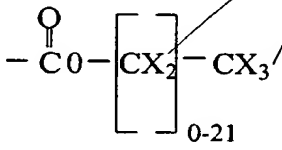
WHAT IS CLAIMED IS:

1. A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder an effective amount of a compound of Formula I:



(I)

wherein:
R_1 is OH, $NHCOCH_3$, or NH_2,
R_2 is H, CO_2H, or



(I)

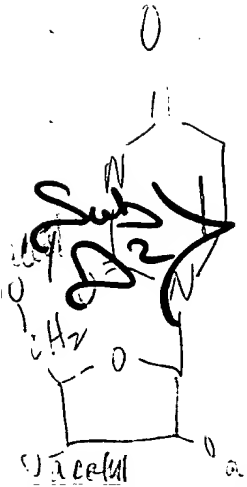
wherein:

X is C_1-C_{22} alkyl, C_1-C_{22} alkenyl or C_1-C_{22} alkynyl, with substituents selected from the group consisting of H, C_{1-3} alkyl, OH, NH_2, and halogen, or wherein X is H,

R_3, R_4, and R_5 are, independently, optionally substituted C_1-C_{22} alkyl carbonyl, with substituents selected from the group consisting of C_{1-3} alkyl, OH, NH_2, halogen, and H, wherein at least one of R_3, R_4, and R_5 is not H,

thereby treating the disorder.

2. A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder, an effective amount of 2', 3', 5'-tri-*O*- acetyl-1-β-D-uridine.



3. A method according to claim 1 or claim 2, wherein the optionally substituted alkyl carbonyl is unbranched and has in the range of about 5 to 22 carbons.

4. A method according to claim 1, wherein the alkyl carbonyl is a carbonyl derivative of an amino acid selected from the group consisting of glycine, L-forms of alanine, valine, leucine, isoleucine, tyrosine, proline, hydroxyproline, serine, threonine, cystine, cysteine, aspartic acid, glutamic acid, arginine, lysine, histidine, carnitine, and ornithine.

5. A method according to claim 1, wherein the alkyl carbonyl is a carbonyl derivative of a dicarboxylic acid having in the range of about 3 to 22 carbons.

6. A method according to claim 1, wherein the mitochondrial disorder comprises a mutation in mitochondrial or nuclear DNA.

7. A method according to claim 1 or claim 2, wherein the mitochondrial disorder is selected from the group consisting of:

Huntington's disease,

Amyotrophic lateral sclerosis,

MELAS (Mitrochondrial encephalomyopathy with lactic acidemia and stroke-like episodes),

MERRF (Myoclonus, epilepsy, and myopathy with ragged red fibers),

NARP/MILS (Neurogenic muscular weakness, ataxia, retinitis pigmentosa/Maternally inherited Leigh syndrome),

LHON (Lebers hereditary optic neuropathy) "Mitochondrial blindness",

KSS (Kearns-Sayre Syndrome),

PMPS (Pearson Marrow-Pancreas Syndrome),

CPEO (Chronic progressive external opthalmoplegia),

Leigh syndrome,

Alpers syndrome,

Multiple mtDNA deletion syndrome,

MtDNA depletion syndrome,

Complex I deficiency,

Complex II (SDH) deficiency,

Complex III deficiency, Cytochrome c oxidase (COX, Complex IV) deficiency,
Complex V deficiency,
Adenine Nucleotide Translocator (ANT) deficiency,
Pyruvate dehydrogenase (PDH) deficiency,
Ethylmalonic aciduria with lactic acidemia,
3-Methyl glutaconic aciduria with lactic acidemia,
Refractory epilepsy with declines during infection,
Asperger syndrome with declines during infection,
Autism with declines during infection,
Attention deficit hyperactivity disorder (ADHD),
Cerebral palsy with declines during infection,
Dyslexia with declines during infection, materially inherited thrombocytopenia and leukemia syndrome,
MNGIE (Mitrochondrial myopathy, peripheral and autonomic neuropathy, gastrointestinal dysfunction, and epilepsy),
MARIAHS syndrome (Mitrochondrial ataxia, recurrent infections, aphasia, hypouricemia/hypomyelination, seizures, and dicarboxylic aciduria),
ND6 dystonia,
Cyclic vomiting syndrome with declines during infection,
3-Hydroxy isobutryic aciduria with lactic acidemia,
Diabetes mellitus with lactic acidemia, — hormonal
Uridine responsive neurologic syndrome (URNS),
Familial Bilateral Striatal Necrosis (FBSN),
Aminoglycoside-associated deafness,
Dilated cardiomyopathy,
Splenic Lymphoma,
Wolfram syndrome,
Multiple mitrochondrial DNA deletion syndromes, and
Renal Tubular Acidosis/Diabetes/Ataxis syndrome.

8. A method according to claim 1 or claim 2, wherein the mitochondrial disorder is a deficiency of cardiolipin.

9. A method according to claim 1 or claim 2, wherein the mitochondrial disorder comprises a deficiency in a pyrimidine synthetic pathway.

10. A method according to claim 9, wherein the deficiency in a pyrimidine synthetic pathway is the uridine synthetic pathway.

11. A method according to claim 9, wherein the deficiency comprises reduced expression and/or activity of an enzyme in the pyrimidine synthetic pathway.

12. A method according to claim 11, wherein the enzyme is selected from the group consisting of dihydroorotate dehydrogenase (DHOD), and uridine monophosphate synthetase (UMPS).

13. A method according to claim 1 or claim 2, wherein the mitochondrial disorder results in lower than normal uridine levels.

14. A method according to claim 1 or claim 2, wherein the mitochondrial disorder is the result of prior or concurrent administration of a pharmaceutical agent.

15. A method according to claim 14, wherein the pharmaceutical agent is a reverse transcriptase inhibitor, a protease inhibitor or an inhibitor of DHOD.

16. A method according to claim 15, wherein the reverse transcriptase inhibitor is Azidothymidine (AZT), Stavudine (D4T), Zalcitabine (ddC), Didanosine (DDI) or Fluoroiodoarauracil (FIAU).

17. A method according to claim 15, wherein the protease inhibitor is Ritonavir, Indinavir, Saquinavir or Nelfinavir.

18. A method according to claim 14, wherein the DHOD inhibitor is Leflunomide or Brequinar.

19. A method according to claim 1 or claim 2, further comprising the administration of one or more co-factors, vitamins, or mixtures of two or more thereof.

20. A method according to claim 19, wherein the co-factor is one or both of Coenzyme Q or calcium pyruvate.

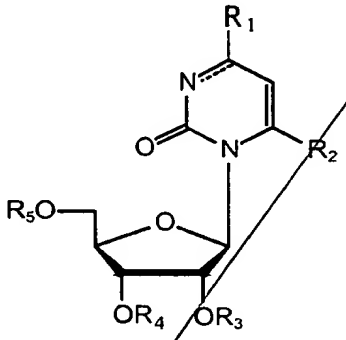
21. A method according to claim 19, wherein the vitamin is selected from the group consisting of thiamine (B1), riboflavin (B2), niacin (B3), pyridoxine (B6), folate, cyanocobalamine (B12), biotin, and pantothenic acid.

22. A method according to claim 1, wherein the compound of Formula (I) is administered in a daily dosage in the range of about 0.5 g/m^2 to 20 g/m^2.

23. A method according to claim 1, wherein the compound of Formula (I) is administered in a daily dosage in the range of about 2 g/m^2 to 10 g/m^2.

24. A method according to claim 1, wherein the compound of Formula (I) is administered in a daily dosage of about 6.0 g/m^2.

25. A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject in need thereof an effective amount of a compound of Formula I:



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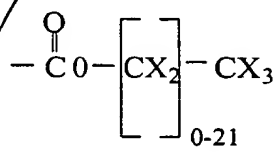

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thereby treating the disorder.

26. A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder, an effective amount of triacetyluridine.

27. A method according to claim 25 and claim 26, wherein the symptoms are renal tubular acidosis (RTA), impaired eyesight, dementia, seizures, cardiomyopathy, skeletal myopathy, peripheral myopathy or autonomic myopathy.